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Proximal Splenorenal Shunts for Extrahepatic Portal Venous Obstruction in Children

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Objective
The results of proximal splenorenal shunts done in children with extrahepatic portal venous obstruction were evaluated.

Summary Background Data
Extrahepatic portal venous obstruction, a common cause of portal hypertension in children in India, is being treated increasingly by endoscopic sclerotherapy instead of by proximal splenorenal shunt. It is believed that surgery (or the operation) carries high mortality and rebleeding rates and is followed by portosystemic encephalopathy and postsplenectomy sepsis. However, a proximal splenorenal shunt is a definitive procedure that may be more suitable for children, particularly those who have limited access to medical facilities and safe blood transfusion.

Methods
Between 1976 and 1992, the authors performed 160 splenorenal shunts in children. Twenty were emergency procedures for uncontrollable bleeding and 140 were elective procedures — 102 for recurrent bleeding and 38 for hypersplenism.

Results
The overall operative mortality rate was 1.9%—10% (3/160–2/20) after emergency operations and 0.7% (1/140) after elective operations. Rebleeding occurred in 17 patients (11%), and pneumococcal meningitis developed in 1 patient who recovered later. Encephalopathy did not develop in any patient. Four patients died in the follow-up period — two of rebleeding, one of chronic renal failure and a subphrenic abscess, and one of unknown causes. The 15-year survival rate by life table analysis was 95%.

Conclusions
A proximal splenorenal shunt, a one-time procedure with a low mortality rate and good long-term results, is an effective treatment for children in India with extrahepatic portal venous obstruction.

Extrahepatic portal venous obstruction (EHO), the most common cause of upper gastrointestinal hemorrhage in children,[1,2] is being treated increasingly by endoscopic sclerotherapy (EST) instead of by surgery. The main reasons for this change are that EST is easy to perform, atraumatic, and effective in preventing bleeding, once the esophageal varices are obliterated. Surgery is favored less because it is thought to be accompanied by

major blood loss and a high mortality rate.[3,4] In the long run, the risks of shunt thrombosis, rebleeding, and portosystemic encephalopathy occurrence are high. Additionally, splenectomy, especially in children, renders patients vulnerable to overwhelming infection.

A splenorenal shunt is a one-time procedure that prevents variceal bleeding.[1] Thus, it might be more appropriate for patients with limited access to blood transfusion facilities than EST, which requires repeated hospital visits. We have been performing this operation since 1976, whenever possible, on all children with EHO referred to us, and present the results of a prospective study.

PATIENTS AND METHODS

Between January 1976 and December 1992, we were referred 192 children (under age 14) with EHO for operation. In 160 children, we performed proximal splenorenal shunts. The other 32 underwent esophagogastric devascularization (27) or mesocaval shunts (5) because previously they had a splenectomy (9), the main splenic vein was not present (19), or the main splenic vein was injured during mobilization for the splenorenal shunt (4).

There were 120 boys and 40 girls (M:F=3:1), whose mean (± SD) age was 10.6 ± 3 years (range, 2 to 14 years). Twenty patients had emergency operations for massive bleeding, uncontrolled by a Sengstaken-Blakemore tube (before 1985) or EST. One hundred forty patients had elective procedures — 102 patients had repeated bouts of massive bleeding (on 3 ± 2 occasions), requiring an average 2100-mL blood transfusion (range, 0 to 16,000 mL), and 38 patients had large spleens that were painful and accompanied by hypersplenism. Only six patients gave a history of umbilical sepsis at birth. Fifteen patients had ascites during a bleeding episode; in only 1 patient (aged 4 years) was it present without bleeding. A history of jaundice was elicited in 25 patients. In 12 patients, jaundice was probably due to viral hepatitis; in 3 patients, it was due to Gilbert's disease, and in 1 patient, it occurred during the neonatal period. In 9 patients, the cause was not clear. One child with chronic renal failure and EHO, had an emergency operation for uncontrolled bleeding.

Preoperative investigations included a routine hematologic examination, liver function tests, and, occasionally, liver biopsy. To evaluate whether the splenic vein was patent, we performed contrast splenoportography or ultrasound examination.

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The operative procedure was performed using a thoracoabdominal incision through the left eighth intercostal space. The spleen was removed, and 4 cm of the splenic vein were mobilized off the pancreas carefully so that it curved gently toward the renal vein. The patient was heparinized, and a splenorenal anastomosis was performed using a continuous 6-0 prolene suture with a "growth factor."[5] If the splenic vein was less than 6 mm in diameter, the anterior layer was closed with 6-0 interrupted prolene sutures.

The patients were included in follow-up postoperatively at 3-month intervals for the first year, 6-month intervals for the second year, and annually thereafter. Those who defaulted were traced by letter and via their local doctors and village health workers. They were not prescribed antipneumococcal vaccine or long-term antibiotic treatment. A follow-up rate of 91% (146/160) was achieved between 1 and 15 years.

RESULTS

Investigations

The mean hemoglobin level was 8.8 ± 2.9 g/dL, the total leukocyte count was 4920 ± 2600 per mm^3, and platelet count was 155,000 ± 88,700 per mm^3. Results of the liver function tests were normal, except for a low level of serum albumin of 3.1 ± 2.9 g/dL and a high alkaline phosphatase level of 23.9 ± 19.8 KAU (normal: 3–13 KAU). After the operation, the mean hemoglobin level was 10.7 ± 1.8 g/dL and the mean serum albumin level was 4.2 ± 0.4 g/dL.

Operation

The average blood loss during the operation was 735 ± 315 mL. The procedure took 3.5 ± 0.5 hours, and the removed spleen weighed 604 ± 442 g. The patient usually was discharged from the hospital on the sixth postoperative day. Two of 20 patients (10%) died after emergency operations and one of 140 (0.7%) died after elective procedures.

Rebleeding

Rebleeding occurred in 17 patients (11%): eight during the first year after surgery, two in the second year, four in the third year, and three others after 4, 5, and 7 years. Eight of these patients were given EST and seven underwent mesocaval shunts. Five of patients in the latter group have not had any rebleeding. Two patients died at home following rebleeding because they could not reach a hospital.

Table 1. DETAILS OF THE FOUR PATIENTS WHO DIED AFTER DISCHARGE

Patient No.	Age	Sex	Cause of Death	Interval After Operation (mo)
1	13	M	Chronic renal failure, subphrenic abscess	3
2	12	M	Unknown	15
3	5	M	Rebleeding	26
4	13	F	Rebleeding	6

Encephalopathy

No patient had encephalopathy after operation.

Infective Complications

Pneumococcal meningitis developed in one child; he recovered after antibiotic treatment. Thirty-eight patients had recurrent attacks of malaria, which were controlled by a single tablet of chloroquine twice weekly.

Long-Term Survival

Life table analysis showed a 95%, 15-year survival. Four patients died at varying intervals after operation due to causes shown in Table 1.

DISCUSSION

Our experience indicates that for children with EHO, the central splenorenal shunt is an effective and safe procedure. The operation could be completed within 4 hours, with less than 1 L of blood loss. It carried an overall mortality rate (including emergency procedures) of 1.9%. There was no long-term encephalopathy, the incidence of postsplenectomy infection was low, and, although the rebleeding rate was 11%, most of these children were managed effectively by EST or by a mesocaval shunt. The previously held belief was that children with EHO should have their bleeding episodes managed conservatively until they grow out of the disease in adolescence by the formation of more extensive collateral circulation. This is probably no longer valid. An individual bleeding episode in EHO, even in western countries, carries a mortality of up to 31%. In the developing world, there is the added danger that diseases, such as acquired immunodeficiency syndrome and hepatitis, could be transmitted through repeated blood transfusions. Further, these children and their parents may be put under considerable emotional and financial stress from numerous hospital admissions and long periods of school absence. These children may not grow as fast as their siblings.[6] The experience with conservative management in India has been disastrous; when one surgeon attempted to trace 25 children with EHO who had refused operation 5 years previously, all of them had died.[7] Thus, active intervention is probably more appropriate. There is little experience with the use of β-blocking drugs[8] in EHO, and the main choice lies between EST and portosystemic shunt surgery.

Endoscopic sclerotherapy is advantageous, as it is the less traumatic procedure and therefore is more widely available, but in EHO, it requires an average of eight[9] sittings to obliterate varices over 8[10] to 24[11] months. Until obliteration is achieved, children remain at risk from bleeding. Complications are more frequent in children and include esophageal stricture (16%),[10] ulceration (29%),[10] and perforation (10.5%).[12] Further, because the portal system is not decompressed, these patients are not only at a 16%[9] risk from variceal recurrence in the esophagus, but 11%[12] have new varices develop in the stomach and mortality rates of up to 5%[12] from bleeding.

A number of different operations, including decongestive procedures and selective shunts, have been performed for children with EHO with low mortality and rebleeding rates. However, our experience and that of others with non-shunt procedures in other situations[13] has not been successful because of rebleeding rates of 30% to 40%, again possibly because the hypertensive portal system has not been decompressed. The main advantage of a selective shunt, such as the distal splenorenal or Warren shunt, is that it is followed by a lower incidence of encephalopathy than total shunts, at least in the short term. This may not be relevant in children with EHO because post-shunt encephalopathy probably does not occur in them.[14] Reports of postoperative encephalopathy in this condition have been from patients who have had EHO secondary to other diseases;[15] reports also came from cases where the encephalopathy included children needing a psychiatric consultation,[16] some of whom had not undergone portosystemic shunting. The Warren shunt also leaves behind the large spleen, which often causes discomfort and hypersplenism. These features may not be reversed after splenic vein decompression.[17]

The high rates of rebleeding after splenorenal shunts reported in other series have occurred when the procedures have been performed by many different surgeons, each of whom have done a small number of operations.[18] Clatworthy and Boles, in their seminal article,[19] reported on 11 patients and found blocked shunts had developed in 4 patients over a variable period. However, when larger experiences are reported from single units, the rebleeding rates range from 5%[20] to 10%.[21] Even if rebleed-

ing does occur, this can be controlled in most instances by EST or by a more proximal anastomosis, such as a mesocaval shunt. An added advantage of operation is the rise in the hemoglobin level by 1.9 g/dL and in the serum albumin level by 2.1 g/dL, possibly because of the reduction of bleeding episodes and the removal of the enlarged spleen.

We cannot explain why we have encountered such a low rate of postsplenectomy infection compared with western countries. We did not use any prophylactic measures because these would be expensive for our patients and we anticipated that compliance would be poor. Others in third world countries, such as Mexico, have had experiences similar to ours;[22] one explanation might be that children living in poor hygienic conditions are immune to overwhelming infection after recurrent attacks of gastrointestinal infections.

The proximal splenorenal shunt for children is a safe, effective, one-time procedure that restores nine of ten children to a normal life and probably normal span. It is the ideal treatment for this disease in children of third world countries, especially in areas where access to sophisticated medical care is limited.

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